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AUTHOR(S):

DAITOH, NOBUYOSHI; TATSUTA, NORIKAZU;
AOSHIMA, MINORU; MATSUDA, MITSUHIKO;
YAMADA, KINYA; HIKASA, YORINORI

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Cor Triatriatum : Diagnosis before Operation and Successful Surgical Treatment

NOBUYOSHI DAITOH, NORIKAZU TATSUTA, MINORU AOSHIMA
MITSUHIKO MATSUDA, KINYA YAMADA, and YORINORI HIKASA

The 2nd Department of Surgery, Kyoto University, School of Medicine.

(Director : Prof. Dr. YORINORI HIKASA)

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We have successfully operated on a 5-year-old boy who had been followed for 4 years with a prior diagnosis of pulmonary hypertension secondary to ventricular septal defect or mitral stenosis. Having used the techniques of catheterization, angiography and echocardiography, cor triatriatum was strongly suspected. The precise anatomy was ascertained during the operation and the lesion was treated successfully by excising the abnormal septum. From the morphogenetic point of view, we obtained some unusual findings associated with the abnormal septum and the interatrial septum.

Case Report

A 5-year-old boy with cardiac failure was admitted to Kyoto University Hospital on Jan. 7, 1975.

A normal pregnancy and delivery with a birth weight of 3.8kg occurred. At the age of 8 months, a cardiac murmur was detected and a ventricular septal defect was suspected. The patient was admitted to a local hospital at the age of 3 years because of initial severe symptoms of high fever and vomiting, and a diagnosis of bacterial endocarditis followed. Mitral stenosis with pulmonary hypertension was suggested after the first cardiac catheterization in Oct. 1974. Fatigability and exertion dyspnea remained unchanged after his discharge. Shortness of breath on exertion had steadily progressed until to the point whereby the patient was unable to walk around his bed and hoarseness appeared.

Physical examination on admission to Kyoto University Hospital revealed an emaciated boy with orthopnea even when digitalis and diuretics were used. The positive findings were a prominent midsternal buldge with a right ventricular impulse, normal first heart sound, loud second heart sound with narrow splitting in the pulmonary area, a grade 2/6 rumbling diastolic murmur at the apex and a grade 1/6 ejection murmur in the aortic valve area.

Chest roentgenograms demonstrated moderate cardiomegaly, a prominence of the pulmonary artery, a double right cardiac contour and signs of marked pulmonary venous

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Present Address : The 2nd Department of Surgery, Kyoto University, School of Medicine.
Sakyo-ku, Kyoto, Japan. 〒 606

congestion. Electrocardiographic findings of right axis deviation, left atrial enlargement and right ventricular hypertrophy with "strain" were present.

The echocardiographic study demonstrated the presence of an abnormal irregular echo in an extremely enlarged left atrium, particularly during ventricular diastole. Despite the diminished slope of the echo from the anterior leaflet of the mitral valve, the echoes from the leaflets showed no thickening.

The results of cardiac catheterization via the saphenous vein and the femoral artery are listed in Table 1. There was no evidence of a shunt at any level, and elevated pulmonary artery pressure was shown. Pulmonary artery wedge pressure was impossible to obtain and the patent

Table 1. Catheterization

	Pre-Op.	Post-Op.
RA	$\bar{3}$	$\bar{2}$
IVC	$\bar{2}$	$\bar{2}$
RV	85/-3	53/-4,
m-PA	88/59, $\bar{70}$	51/24, $\bar{38}$
r-PA		53/22, $\bar{38}$
r-PC		$\bar{9}$
LV	108/-4	

foramen ovale was not identified.

Two preoperative angiograms were obtained. The first injection of the contrast medium was made into the right ventricle.

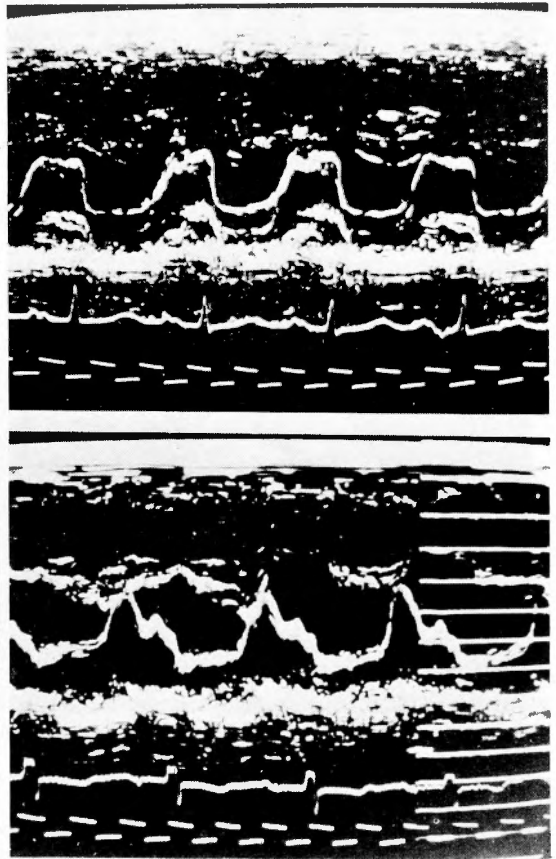


Fig. 2. (above) Preoperative echocardiogram. (below) Postoperative echocardiogram. Abnormal echo from the diaphragm is no longer apparent and the diastolic descent rate is normal.



Fig. 1. Preoperative angiogram. Only the accessory left atrial chamber is opacified.

The pulmonary circulation time was delayed. One extremely enlarged chamber was well delineated having a vertical membranous septum along its left border. The second injection was made into the left ventricle through the aortic valve. A small true left atrium was opacified due to mitral regurgitation. Another cine-angiogram revealed that the left appendage was located downstream of the abnormal septum.

From this evidence of abnormal membrane, the diagnosis was cor triatriatum, not mitral stenosis.

Corrective surgery was performed on Feb. 24, 1975, with the aid of cardiopulmonary bypass. Following a transverse incision in the right atrium, there was no evidence of an atrial septal defect or an patent foramen ovale. The thick atrial septum was opened using the DuBost technique. As expected from the preoperative findings, the left atrium was divided into two chambers by an abnormal septum with a small opening, 10 mm in diameter. All four pulmonary veins drained into the enlarged accessory chamber, and the left atrial appendage communicated with the small true left atrium just below the membranous septum. The membrane was completely excised, reducing the two atrial components to one single cavity and revealing an entire mitral valve with an orifice, 33 mm in diameter. No manipulation of the competent mitral valve was made.

The postoperative recovery was uncomplicated. The patient became asymptomatic, hoarseness disappeared, and he began a physically vigorous life. Postoperative right heart catheterization was carried out on Jun. 10, 1975. Although pulmonary artery pressure (mean 38 mmHg) remained slightly elevated, wedge pressure had returned to normal (9

mmHg). The right ventricular angiogram revealed a markedly smaller left atrial cavity. Echocardiography showed no abnormal echo from the left atrium, and normal movement of the leaflets of mitral valve were demonstrated.

Comment

Cor triatriatum is a rare congenital cardiac anomaly in which the left atrium is divided into two chambers by an abnormal septum with a small opening. When not associated with other anomalies, this disease is hemodynamically analogous

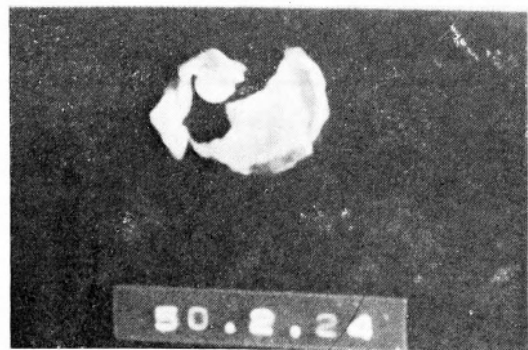


Fig. 3. Abnormal diaphragm with an opening, 10 mm in diameter.

to mitral stenosis and causes pulmonary venous obstruction of various degrees of severity depending on the size of the opening between the accessory chamber (high pressure chamber) and the true left atrium (low pressure chamber). When the lesion is associated with other cardiac anomalies, the clinical findings are complicated and preoperative diagnosis is difficult to make. The presence of this lesion may be not recognized even during the

surgical procedures.

In recent years, increasing attention has been paid to cor triatriatum, and reports of successful surgical correction have been growing in number. The case reported here was a classical form of this lesion with no additional cardiac anomalies. The presence of an abnormal memberanous septum was demonstrated preoperatively by angiogram and echocardiogram.

The noninvasive echocardiographic technique has been found to be useful as a complementary method for the investigation of congenital heart disease. It can be easily used on seriously ill infants.

The abnormal echoes from within the left atrium come from the following lesions : 1) left atrial clots, 2) left atrial tumors, 3) abnormal membranous structures.

Most clots are small and occur in the vicinity of the left atrial appendage, an area virtually impossible to examine with current echocardiographic techniques. Occasionally, a very large left atrial clot may cause several linear echoes along the posterior left atrial wall.

The most common atrial tumor is myxoma, which is pedunculated and arises via a stalk from the interatrial septum. Left atrial myxomas do not have the same echocardiographic characteristics. The tumor may appear as a group of echoes posterior to the anterior leaflet of mitral valve. This cloud of echoes originates from the tumor which falls through the mitral orifice behind the anterior mitral leaflet. One must have sufficient gain to obtain the echo, but a high gain setting may also cause confusion thickened and stenotic posterior leaflet with a left atrial tumor. It is suggested that the easiest way to eliminate confusion is to scan toward the left atrium. In patients with a true left atrial myxoma, the tumor echoes would persist and possibly become even prominent as the ultrasonic beam entered the area of the left atrium¹⁾.

Abnormal membranous structures which represent irregular linear or laminar echoes in the left atrium come from cor triatriatum and supravulvar stenosing ring. Few echocardiographic findings of these two lesions have been reported²⁾³⁾. Some investigators pointed out that the pattern of movement of the abnormal echo from the membrane within the left atrium was characterized by a slight movement during ventricular systole and a more pronounced movement during ventricular diastole, especially during atrial systole.

Echocardiographic technique is not sufficient for differential diagnosis of cor triatriatum and supravulvar stenosing ring. Angiocardiography should be done in order to determine whether the left appendage communicates with the upper or lower chamber of the left atrium⁴⁾.

In the current case, linear oscillating echoes were obtained in diastole, and the diastolic descent rate of the anterior mitral leaflet was remarkably decreased without thickening. After surgical treatment, the abnormal echo disappeared and valvular leaflets displayed normal movements with a characteristic A wave. It also shows that after resection of the abnormal septum, blood flow through the mitral orifice increased.

There have been three principal hypotheses concerning the morphogenesis of cor triatriatum : 1) abnormal growth of septum primum, i.e. malseptation;⁵⁾⁶⁾ 2) failure of incorporation of the common pulmonary vein into the the left atrium, i.e. malincorporation;⁷⁾⁸⁾ and 3) entrapment hypothesis.

In the entrapment hypothesis, VAN PRAAGH and CORSINI⁹⁾ pointed out that the subdividing diaphragm (abnormal septum) was composed of the wall of the common pulmonary vein dorsally and the wall of the primitive left atrium ventrally. From their hypothesis the foramen ovale should be open to the true left atrium. They thought earlier reports of the foramen ovale opening into the dorsal (accessory) chamber were misinterpretations.

From this hypothesis, fossa ovalis would also be expected to lie against the wall of the true, rather than the accessory left atrial chamber. In the majority of previously reported cases, certainly, the fossa ovalis lay between the right atrium and the true left atrium.

In contrast, however, fossa ovalis was definitely located between the right atrium and the accessory chamber in our case. Some reports gave attention to the location of the fossa ovalis, and several cases were present in which it was found against the wall of the accessory chamber¹⁰⁾¹¹⁾.

Although the entrapment hypothesis is becoming influential in describing the morphogenesis of cor triatriatum especially through the prominent works by VAN PRAAGH and CORSINI, it seems the malseptation theory is more accurate in explaining the case discussed in this paper.

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和文抄録

三心房症：術前診断のできた1治験例

京都大学医学部外科学教室第2講座（主任：日笠頼則教授）

大頭信義，龍田憲和，青嶋 実

松田光彦，山田公弥，日笠頼則

三心房症は発生頻度の極めて低い心奇形であるが、しばしば合併奇形を伴うため術前診断が困難であり、術中においてすら見逃されることがある。近年、本症への関心が高まり、心臓カテーテル検査、心血管造影、超音波検査による術前診断及び手術成功例の報告も次第に増加している。

患者は5才の男児で、生後8ヶ月に心雑音を指摘され、心室中隔欠損症を疑われた。3才8ヶ月の頃より心不全をくり返し、他の医療施設にて心臓カテーテル検査を行い、僧帽弁狭窄症の疑いと診断された。

当院における術前検査により三心房症を強く疑われ、1975年2月開心術を施行した。

左心房は異常隔壁によって2つの Chamber に分かれ、その間に径10mmの交通孔があった。僧帽弁は弁尖、腱索ともに異常なく、隔壁を完全に切除して手術

を終えた。

術後経過は良好で、運動能力の著明な改善をみた。3ヶ月半後の心臓カテーテル検査では、術前、70mmHgの平均肺動脈圧は38mmHgとやや高値に留まっているが、楔入圧は9mmHgと正常値を示している、また心血管造影では左房の縮小が明瞭に認められ、超音波検査にても、左房内の異常エコーが消失して、僧帽弁尖がM型の正常な動きをすることが示された。

なお、本症例の特異的な所見は fossa ovalis が accessory (high pressure) chamber に面していた点であり、近年、三心房症の異常隔壁の成因に関して有力視されつつある Van Praagh らの entrapment hypothesis では説明できず、むしろ malseptation theory の方が理解しやすい。